

Chapter 2

COMPREHENSIVE CARE OF HEMOPHILIA

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COMPREHENSIVE CARE

Hemophilia is a rare inherited disorder that is complex to diagnose and to manage. Optimal care, especially for people with severe forms of the disorder, requires more than treatment of acute bleeding.

Prevalence

17 per 100,000 males have hemophilia A
4 per 100,000 males have hemophilia B

**For every 100,000 males,
21 have hemophilia**

Prevalence at birth

25 per 100,000 male births, have hemophilia A
5 per 100,000 male births, have with hemophilia B

**For every 100,000 male births,
30 are born with hemophilia**

Hemophilia should be suspected in people with these symptoms:

- ✓ Easy bruising
- ✓ "Spontaneous" bleeding (i.e., bleeding for no known reason), particularly into the joints, muscles, and soft tissues
- ✓ Excessive bleeding following trauma or surgery

The severity of bleeding manifestations in hemophilia generally correlates with the degree of the clotting factor deficiency.

MILD Hemophilia

Clotting factor level:



- 5-40 IU/dL (5-<40% of normal)
- Rare spontaneous bleeding
- Severe bleeding only with major trauma or surgery

MODERATE Hemophilia

Clotting factor level:



- 1-5 IU/dL (1-5% of normal)
- Occasional spontaneous bleeding
- Prolonged bleeding after trauma or surgery

SEVERE Hemophilia

Clotting factor level:

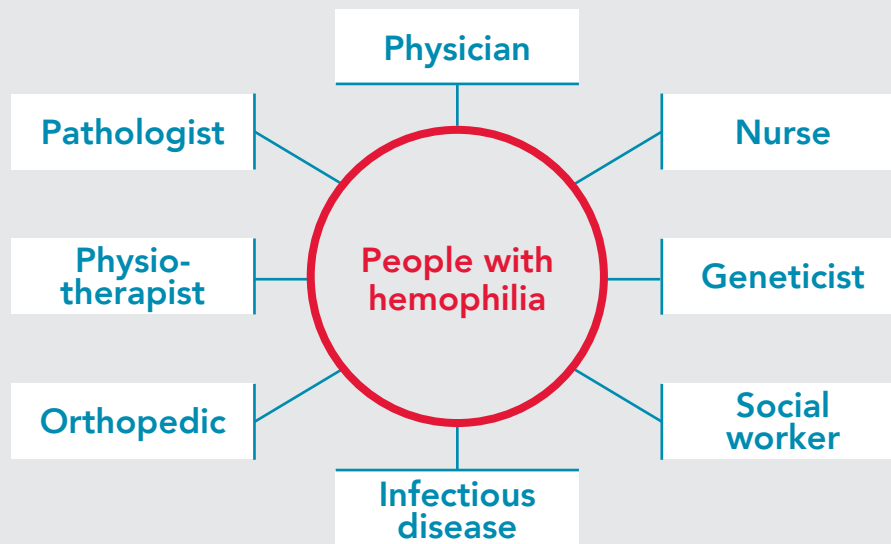


- <1 IU/dL (<1% of normal)
- Spontaneous bleeding into joints or muscles

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MULTIDISCIPLINARY COMPREHENSIVE CARE TEAM

Comprehensive care of hemophilia involves multidisciplinary medical services necessary for the diagnosis, treatment, and management of hemophilia and its complications, delivered by core members of the comprehensive care team at hemophilia treatment centres.



Key Components of Comprehensive Care

- Prevention of bleeding and joint damage
- Prompt management of bleeding episodes, physical therapy and rehabilitation after joint bleeds
- Pain management
- Management of musculoskeletal complications
- Prevention and management of inhibitors and other co-morbidities
- Dental care
- Quality-of-life assessments and psychosocial support
- Genetic counselling and diagnosis
- Ongoing patient/family caregiver education and support
- Emergency care should be available at all times with availability of lab services and treatment